

R H E U M A T I C E N D O C A R D I T I S

I N C H I L D R E N .

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by

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RHEUMATIC ENDOCARDITIS IN CHILDREN.

The term "Rheumatism" has probably been ascribed by the profession to diseases which are essentially different both pathologically and clinically. To term a condition "rheumatic" is too often to cloak one's ignorance of what the true diagnosis is. In spite of the careful and tedious researches of such men as Church, Triboulet, Still, Poynton, Paine, Singer, etc. our knowledge of what we term "Rheumatism" is still imperfect. The pathology has only of late years been at all accurately worked out. As it is upon this that a true diagnosis of rheumatism hangs, I am desirous of referring to it at some length.

On the other hand it must be confessed that in practice we meet with a class of diseases termed "rheumatic", which on superficial examination appear to be different, but if submitted to a closer analysis are seen to be intimately allied. Few can deny the close connection which exists between rheumatism and chorea. How frequently do we find a tonsillitis the only manifestation of what is really an attack of true rheumatism.

The term "rheumatic diathesis" which has been used by many has certainly some grounds for its adoption. I have myself frequently observed (see cases) different manifestations of rheumatism occurring at different times in the same individual and dissimilar types of the disease in two members of the same family.

These different manifestations of, or allied conditions to rheumatism shew more or less a tendency to the development of Endocarditis. I have thus chosen this subject of study, namely Endocarditis, as a basis from which to refer my observations upon them all. In children these relations are most characteristic and for brevity's sake I have therefore more particularly confined my remarks to them.

During my two years of office as House Surgeon and Physician to the Ingham Infirmary, South Shields and Westoe Dispensary, I have been extremely fortunate in seeing an almost phenomenal number of so-called rheumatic cases. The time at my disposal has naturally been small, and in the hurry of dispensary practice many interesting cases have been allowed to slide without notes being taken. The objects of this paper are simply to give the results of my own observations,

coupled with the study of the literature of competent authorities in this very fertile field of research. The general ideas and theories which these beget in my mind I have tried to place in writing.

I often wonder if the geographical and climatic conditions which are present at South Shields favour the universal predominance of rheumatism amongst the inhabitants. Here we have a seaport town at the mouth of the river Tyne, where bracing though moist sea breezes are very prevalent. The higher parts of the town are healthy, but in the low-lying dwellings of the poor, the atmosphere is very cold and damp. Such conditions, associated with the overcrowding which is present, make a fertile soil for the ravages not only of rheumatism but also of Phthisis.

Rheumatic endocarditis as met with in children differs in no essential respects from that which is met with in adults. It is an inflammation of the lining membrane of the heart, due to its invasion by what has now been almost conclusively proved to be the diplococcus of rheumatism. The widespread character of the disease is only too well known, but I am of opinion that it would prove to be still more common in its

milder forms, were a more systematic examination of the heart made in children. In its earlier forms, associated as it may be with chorea, tonsillitis, growing pains, it is too often missed, and it is only when the subsequent symptoms of heart disease appear that the physician recognises the presence of a preliminary endocarditis. When examining the heart previous to the administration of an anaesthetic in over 300 cases, I have frequently detected a systolic murmur. That a mere systolic murmur is not necessarily organic has been frequently shewn, but a certain percentage would probably be so. I regret not having inquired in all cases for a previous history of some manifestation of rheumatism, and it would be interesting to follow the subsequent history of such cases.

Not only, then, is this disease a very common one in childhood, but is also a very serious one. It is distressing to anticipate the disastrous effects of such a small pathological entity in the subsequent horrors of heart disease which make our patient a life-long invalid or bring him to a premature grave.

It should, therefore, be our endeavour to make an early diagnosis under whatever mask of

other symptoms its insidious onset takes. As it is only in these early stages that our efforts are of really any avail in its cure, we should study these masking symptoms, and carefully examine the heart if there be any suspicion that they are of rheumatic origin. For example, it is only rarely that choreic movements, growing pains, or tonsillitis are of danger to the patient, but their known tendency to an associated endocarditis should make the physician careful.

BACTERIOLOGY.

The bacteriology of rheumatic endocarditis involves the study of the bacteriology of rheumatism in general. There is still a great difference of opinion amongst competent observers as to the cause of the disease. At the present time two main theories hold the field.

1. That it is due to a specific diplococcus.
2. That it is an attenuated pyaemia due to infection with various pus forming cocci.

The first of these seems to have the most adherents. Triboulet and Coyon, Wasserman, Poynton and Paine, have independently of one another isolated diplococci. These diplococci, though differing in some minor respects, appear to

be identical and the cause of the disease. I am indebted to Dr W. E. Marshall for slides of this micro-organism, where the diplococcal nature of the organism is clearly shewn. As described by Poynton and Paine (Encyclopaedia Medica) the micrococcus has the following characteristics. It is minute and essentially a diplococcus, each element of which is .5 m in diameter. On solid media it grows in staphylococcal masses, in liquid media in streptococcal chains, each element of which is a diplococcus. We found the most favourable culture medium was blood agar or milk and bouillon slightly acidified with Lactic acid. It is present in the local lesions and in the blood and urine of the living patients suffering from Pericarditis. Injected intravenously into rabbits it reproduces the disease and can be isolated from the local lesions. The organism is rapidly destroyed at the sites of the local lesions in man and this fact together with its minute size makes it difficult to find in the tissues. We have succeeded in demonstrating the diplococcus in the nodules and in the synovial membrane of the joints and have isolated it from both these structures and from most of the rheumatic lesions, (including the heart valves). It is usually present in the exudations, though in very acute and clear effusions it may be absent or present only in small numbers.

Who?

In suitable media it lives for long periods and keeps in some measure its virulence. It stains well with the aniline dyes, but in the tissues does not retain the gentian violet used in Gram's method with tenacity. The diplococcus certainly gains access through the tonsil and may also do so through paths as yet unknown.

Dr Beattie in a communication (B.M.J. Dec. 3rd 1904) has amply confirmed these results. He states that intra-~~venous~~^{ven} injections of the micrococcus in animals produce endocarditis, polyarthrititis and Chorea. He has not seen suppuration follow injections. The micrococcus differs essentially from the streptococci not only as regards the results of inoculation but also in the cultural characters and vitality of the organism, as markedly as the pneumococci are distinguishable from them.

Walker considers the diplococcus of Paynton and Paine to be indistinguishable from the original streptococcus. It differs, however, in growing abundantly on the filtered culture fluid of that micro-organism (Pract. Feb. 1903).

In a later communication Walker and Ryffel state that this so-called streptococcus or diplococcus produces in a blood agar culture the colour

change characteristic of the pneumococcus and the influenza bacillus, and that it has a haemolytic action on red blood corpuscles greater than any other streptococcus, and that it produces considerable quantities of Formic Acid and another acid of the fatty series. Formic acid is also stated to be found in the bodies of the micro-organisms (Pract. Sept. 19th 1903).

In connection with this production of Formic acid, I think it is interesting to note a phenomenon which has been described and noticed by myself as a point in the differential diagnosis between gonorrhoeal and true rheumatism. If a bee sting a person suffering or who has suffered from true rheumatism no red areola is produced. On the other hand the subject of gonorrhoeal rheumatism exhibits the typical red poisoned wound of the insect. Now it is known that one of the chief constituents of bee's venom is Formic acid and I suggest that the individual suffering from true rheumatism had developed a certain immunity to this form of toxin as an explanation of the phenomenon. With regard to the old lactic^{acid} theory, it is interesting to note a case which occurred in the practice of one of my colleagues. He was administering Lactic acid to a child for some complaint when it suddenly developed

a multiple arthritis. The drug was stopped and the arthritis subsided. On restarting the drug, the arthritis again developed. I did not feel justified in repeating the experiment.

The great propounder of the second theory is Singer who maintains that rheumatism is an attenuated pyaemia due to infection with various pus-forming cocci. He has demonstrated the streptococcus in the tonsils and blood of patients suffering from rheumatic fever. The constancy of the disease is too great to be so regarded until several specifically different pus-forming micro-cocci are proved conclusively to cause rheumatic fever.

With regard to rheumatic endocarditis, tonsillitis, arthritis, pericarditis, etc., we must consider them as local manifestations of a general disease. In this way I compare rheumatism with pneumonia which is also a general disease, but which may shew local lesions in the lungs, joints, meninges. In diphtheria also we have a general disease with a local lesion in the throat but with toxins circulating throughout the blood. In these two diseases specific micro-organisms have been isolated and accepted by nearly everyone as the cause of the diseases. Now if the throat of a diphtheria patient be examined, not only will you find Loeffler's Bacillus, but a large number of streptococci also.

In like manner in the endocarditis which may follow pneumonia, not only do we find the Fraenkel's pneumococcus but also streptococci and other micro-organisms. In fact, in both these diseases, we have a specific micro-organism associated with other micro-organisms chiefly streptococci which evidently play some part in the disease. I suggest a similar condition might be present in rheumatism and this would in some measure reconcile the two opposing theories. It may be that certain forms of soil, temperature, surrounding media favour the growth of these micro-organisms together. If scarlet fever is due to the streptococcus, its frequent association with rheumatic manifestations could be thus explained.

MORBID ANATOMY.

This diplococcus then of rheumatism travelling in the blood shews a great tendency to attack the serous membrane of the heart, and especially those parts of the membrane which are exposed to friction of their surfaces. For the pathological appearances I am indebted in great measure to Professor Greenfield's Lectures and my notes taken from them. In the endocardium the most characteristic effects of the inflammation are in the production of warty

vegetations. It affects first of all the valve curtains and later the rest of the endocardium. In extra-uterine life it is the left side of the heart which is primarily affected. In intra-uterine life it affects the valves of the right side of the heart. In extra-uterine life the right side of the heart in rare instances shews evidence of invasion. The mitral valve is the most frequently attacked. The aortic comes next in frequency, but its affection without involvement of the mitral also is rare. In my cases the mitral valvulitis was by far the commoner, and I have not seen a pure aortic lesion in children due to rheumatism. Affections of the pulmonary valve are seldom met with.

Let us then consider the morbid appearances on the mitral valve. We first see some swelling of the curtains leading to a beaded translucent appearance at the edge of the valve. There is no great increase of vascularity. On microscopic examination this swelling is seen to be due to an invasion of leucocytes and proliferation of connective tissue cells beneath the endothelium. The connective tissue fibres are separated and some of the endothelial covering lost. The diplococcus is to be found amongst the swollen connective tissue. As the change progresses, the chordae tendinae become swollen, soften and may actually rupture. The

ruptured chordae may become attached to others. The inflammation may continue to spread to the papillary muscle, causing it to become paler and injected with blood. We now get the appearance of the vegetations. At first they are minute projections, translucent and slightly red in colour. They appear along the free margins of contact of the valve curtains, and are thus first seen on the auricular surface. As they increase in size they coalesce, producing an irregular roughness. Coagulation now occurs upon them and this is chiefly due to the deposit of blood plates.

In the aortic valves similar changes are seen, modified by their anatomical appearance. Here the vegetations first appear along the Lunulae and Corpus aurantii on the ventricular surface, this being the line of contact of the valves. Aneurism and rupture of these valves may occur, but it is not nearly so common as in ulcerative endocarditis..

Spread of the inflammation may occur from these valves on to the walls of the auricle and ventricle, but again this is much more frequent in the malignant form.

As the disease abates, if the inflammation is not excessive, there is a possibility of a return of the valve to the original condition. The valves

of the heart, however, are in constant motion and complete rest cannot be given for their repair. Consequently, such a satisfactory result is rare, it being much more common to get some of the following secondary effects developed:-

The first effect on the inflamed tissue is that the swelling is followed by shrinking. The most serious effects of this shrinking are produced on the Aortic valve. Segments are thickened and do not move so well. They do not approximate completely and thus we have incompetence as a result. On the mitral valve the effects of this shrinking are not so serious unless the chordae tendinae are also affected, as there is a greater reserve of valve material. Another most serious result of rheumatic endocarditis is that adhesions develop between the margins of the valves. These adhesions may occur between the chordae tendinae and lower part of the valves or higher up at the bases of the cusps producing a funnel-shaped or button-hole orifice respectively. The button-hole orifice is the more serious condition. These adhesive changes are characteristic of the mitral valve and produce the various forms of stenoses of that orifice. Adhesions may occur at the aortic orifice, but they are much rarer, owing to the great shrinking of cusps and rapidity of blood flow over them.

In rheumatic endocarditis, a small embolus may occasionally occur due to the detachment of one of the small vegetations.

Calcification is not met with until adult life.

CAUSES.

In what allied diseases of children then do we meet with rheumatic endocarditis?

Rheumatic fever in its acute form is rare in children. The most marked cases have been associated with scarlet fever as hereafter stated. Endocarditis is a frequent concomitant.

Subacute rheumatism and growing pains.

In children the above are very common, the latter being ^{little} thought of by parents. In the so-called cases of idiopathic heart disease in children I have found on enquiry a history of vague pains in the limbs occurring during earlier childhood.

Rheumatic nodules.

Rather curiously I have notes of only one case of this condition. The boy, aged 12, had well marked endocarditis besides recurring arthritis. The endocarditis eventually proved fatal. These nodules rarely occur without the presence also of endocarditis.

Tonsillitis.

That a tonsillitis is a frequent preliminary manifestation of rheumatism has been recognised by clinicians for many years. I have records of five cases which preceded the arthritis by a fortnight or three weeks. In such cases an opportunity is given for aborting an attack of rheumatism with its possible endocarditis. This preliminary symptom is of especial value and significance to us in second attacks of rheumatism. One can then be more confident that anti-rheumatic remedies have averted an attack. I had one marked case of this. In my primary cases of what I considered to be cases of rheumatic tonsillitis and where no arthritis developed, one was bound to admit that perhaps an arthritis might not have developed, had no treatment been adopted.

Chorea.

It is now generally admitted by authorities that chorea is of rheumatic origin. Its production in animals by the injection of the "micrococcus rheumaticus" by Dr Beattie and other pathologists practically proves it to be such. In nearly all my cases of which I have notes, there was a marked family or personal history of rheumatism. The children were of a neurotic type. Endocarditis was of frequent occurrence in those cases which

had not been kept in bed during a previous attack. Several of my cases of endocarditis in their past histories shewed the following sequence, rheumatic fever or subacute rheumatism, chorea, endocarditis. Months or years intervened between each set of symptoms.

Erythema nodosum and other forms of purpura.

The term purpura is a collective one and probably embraces diseases differing widely in their pathology. Erythema nodosum and peliosis rheumatica have probably the same pathology as rheumatism. I saw a large number of cases of erythema nodosum associated with arthritis but in only one of them did I detect definite endocarditis. In two cases of Henock's purpura and one of hemorrhagic purpura of which I have notes, I was unable to say whether or not they were of rheumatic origin. No endocarditis developed.

Scarlet fever.

The frequent appearance of endocarditis and arthritis in this condition has been to me a subject of most interesting study. They usually occur simultaneously with or in succession to the scarlet fever. The points of interest are these:- Is this scarletinous endocarditis and arthritis of rheumatic origin? and if so, is the bacteriology of the two diseases allied? I have notes of three cases of my own and about ten in those of my col-

leagues, besides those in the following table,
kindly collected for me by Dr Marshall.

	Total	Male	Female.
Total number of Scarlet Fever Cases.	70	24	46
Number with simple rheumatism	4	4	0
Number with simple rheumatism plus endocarditis.	1	1	0
Number with simple rheumatism plus cardiac failure.	1	0	1
Total with rheumatic symptoms	6	1	5
Number with heart affection, without definite signs of rheumatism.			
Endocarditis.	1	0	1
Cardiac failure.	7	3	4
Total	8	3	5.

In their experience and my own the reaction of the arthritis to salicylates was marked and would seem to indicate that it was rheumatic and not pyaemic in origin as some have suggested.

The following case of my own shews a striking and significant sequence of diseases. The girl aged about 12, had first of all an attack of chorea. This was followed by scarlet fever. Then rheumatic fever developed and finally when she consulted me *she* was suffering from endocarditis.

SYMPTOMS AND DIAGNOSIS.

The diagnosis of rheumatic endocarditis is by no means easy. It rests on signs and symptoms which are more or less equivocal. In my opinion for practical purposes, the more important duty that devolves upon us, is to diagnose whether or not we are dealing with a rheumatic condition.

If so, then our case must be treated as if on the brink of developing endocarditis, for we have seen the great tendency of the rheumatic poison to attack the valves of the heart. Endocarditis itself in many cases runs quite an insidious course and its certain diagnosis is practically impossible. If we can detect the rheumatic element in the case early, the danger of an endocarditis may be averted.

What then are the points which we should consider? A careful enquiry should be made into the personal and family history for evidence of rheumatism, chorea, growing pains, tonsillitis, etc. The child should be examined for rheumatic nodules, and most important of all, a careful examination of the throat should be made. Bertram Abrahams (Clin. Soc. Trans. 1899) has shewn by statistics the great importance of the recognition of rheumatic sore throat in children. It appears that a tonsillitis is a more frequent accompaniment of rheumatism in certain localities than in others. This

would seem to prove that there are other sources of infection besides the tonsil. In several of my cases, as above stated, the tonsillitis may precede the rheumatism by a fortnight or three weeks, thus giving us ample opportunity of anticipating the onset of rheumatism and possible endocarditis.

There is need of further study with regard to the bacteriology in order to obtain as in diphtheria a bacteriological examination of the throat for the specific diplococcus.

Achalme describes what he calls the pre-articular symptoms of rheumatism and lays stress on their diagnostic importance. Of these the most significant is cardiac irregularity of which he quotes three cases in addition to one previously recorded by Graves. All the patients had a pulse between 50 and 60 which was irregular for from 48 to 62 hours. There was no palpitation and the symptom disappeared when the joints became affected. In each case the attack was mild, a cardiac murmur only being present in one of them. He considers the symptom due to an early invasion of the myocardium by Bacteria (Arch. Gen. de Med. Sept. 1902). In one of my cases this symptom was present but was coincident with the arthritis. I rather regarded the irregularity, therefore, as due to a commencing endocarditis. It disappeared entirely as the patient

became convalescent and no valvular lesion remained.

Having then satisfied ourselves that in all probability we are dealing with a rheumatic case, what are the signs and symptoms which might indicate the possibility of an endocarditis. The following are described by Osler:- The patient as a rule does not complain of any pain or cardiac distress. In a case of acute rheumatism, for example, the symptoms to excite suspicion would be increased rapidity of the heart's action, perhaps slight irregularity and an increase in the fever without aggravation of the joint trouble. That these small vegetations can excite fever is shewn by the study of recurring endocarditis in old sclerotic valves, and which may be associated for weeks at a time by slight fever ranging from 100° to $102\frac{1}{2}^{\circ}$. Palpitation may be a marked feature and is a symptom upon which certain authors lay great stress.

The diagnosis of the condition rests upon physical signs which are notoriously uncertain. The presence of a murmur or roughening of sounds at one or other of the cardiac areas is often regarded as indicative of the existence of endocarditis. This murmur, however, when taken alone is a very

uncertain sign. Associated with the development of a murmur, we may find accentuation of the second sound in the pulmonary area.

TREATMENT.

The question of treatment resolves itself into prophylactic and curative measures but which are in great measure the same. The more important are the prophylactic and in order to carry these out, the importance of an early diagnosis of rheumatism, under whatever form it appears, is clearly shewn. We have discussed at some length the points that should guide us in forming this opinion. In the various conditions which I have annotated as being allied to rheumatism, rheumatic endocarditis may shew itself to a greater or less extent. Some, as chorea, rheumatic fever, are more prone to it than the others, but we must never close our eyes to the possibility of its occurrence in the other conditions. In the forms of treatment which I have adopted, I have followed in great measure those laid down by Caton for the prevention and cure of valvular disease in the course of rheumatic fever. Their use I have extended to the allied conditions of chorea, rheumatic tonsillitis, growing pains, etc. and without being too optimistic, I feel they have

given me satisfactory results.

When a rheumatic case in a child presents itself, the first measure which I insist on is absolute rest in bed, even if no evidence of endocarditis. It has been shewn that by this means the heart's action will be reduced several beats per minute and the utmost possible physiological rest is given to the valve curtains. It is a distressing fact to see so many cases of well-marked chorea walking the streets, when we know the great risk the heart is running. How long they should be kept in bed, will depend greatly on the case, and whether or not there be evidence of commencing endocarditis. Cases of rheumatic fever I keep in bed five or six weeks and longer where there is an endocardial murmur. Chorea cases sometimes require even longer than this. As it is so difficult to be certain that we are dealing with a rheumatic tonsillitis, purpura, growing pain etc., we are scarcely justified in keeping them in bed so long. It is better, however, to err on the side of safety than to allow a ^{Careful} tonsillitis to get up, and develop a rheumatic fever within a fortnight.

Erythema nodosum and peliosis rheumatica in my experience only rarely develop an endocarditis, only one of my cases doing so. I like to keep them in bed ten days to a fortnight. Cases of rheumatism

associated with scarlet fever will require prolonged rest for the scarlet fever and the danger of a complicating nephritis, besides the danger of an endocarditis.

A daily careful examination of the heart should be made to detect any evidence of endocardial mischief. If such appears we must be more stringent than ever in our efforts to obtain physiological rest for the heart. As we could not expect an inflamed rheumatic synovial membrane to resolve were the joint in constant motion, in like manner we cannot expect an inflamed endocardium to return to normal, unless it be given as much rest as possible.

With regard to diet, it should be kept as low as possible, especially if the case be acute, as again less strain is thrown upon the heart. Caton recommends an entirely milk diet and if patients will endure it, it is perhaps best. After a time a little arrowroot and other carbohydrate foods may be allowed. Beef tea, tea, coffee and alcoholic beverages, and in fact all substances which excite the heart should be forbidden. In convalescence diet should be gradually increased, but tentatively if there is still evidence of endocardial trouble.

As regards drugs, there seem to be none which have a direct influence on the inflammatory process.

Salicylates may be given for the associated rheumatism. Sanson recommends the sulphocarbolates. If there be much vascular excitement aconite may be given. If the heart be very irregular, a small dose of digitalis may be administered, but this drug is as a rule contraindicated. Sodium iodide is the drug which Caton found the most satisfactory in producing resolution and absorption of the inflammatory products. This drug I have frequently used in from two to four grain doses for children combined with salicylates, but it is difficult to differentiate the results of the various forms of treatment adopted.

With regard to local measures, I have followed Caton in placing a series of small blisters in the 3rd, 4th, 5th and 6th interspaces just to the left of the sternum. These blisters by their action on the intercostal nerves are hoped to exercise an influence on the heart through the connections which these nerves have by means of the sympathetic with the cardiac plexuses.

Caton has shewn in his small treatise "Prevention and cure of valvular disease of the heart" by statistics and minute observations that when the above measures were adopted, he reduced markedly the percentage of endocardial complications in rheumatic fever. These measures are no doubt

tedious and their stringency difficult to understand by our patients, but if we can lessen the incidence of heart disease, no trouble should be spared. During my two years out-visiting in the dispensary practice of the hospital, I have striven to carry out these measures. It was a difficult problem in the houses of the poor to keep patients so long in bed when to all appearances they were quite well, but except in odd cases, when the objects in view were explained to them, I was able to get the treatment carried out. As a result my experience has served to strengthen me in the opinion that great benefit will accrue from these measures. In chorea, perhaps, I have obtained the most gratifying results. In none of my cases which were submitted to this treatment, and in whom there was not already evidence of valvular trouble from a previous attack, have I been able to say that there remained a permanent valvular lesion.

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